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*Editorial Note*

# What is a ventricle? Morphologic lessons from the Fontan circulation

Robert H. Anderson

In the accompanying report,[1] Murari and colleagues give a detailed analysis of the morphologic findings within the hearts of a large number of patients they chose to correct surgically by creating a circulation in which flow to the lungs is no longer driven by a ventricle. This, of course, is the situation produced by the Fontan procedure and its various modifications. Description of the ventricular mass in such patients produces potential problems since, although the pulmonary circulation does not have the benefit of its own ventricle to provide systolic propulsion, it is unusual for the patients treated in this fashion to have a solitary ventricular chamber. Indeed, I was involved in a very similar study to this one when I spent a period in Melbourne in 1992, working with Jim Wilkinson and his colleagues. When we analyzed the ventricular morphology of the patients submitted to the Fontan procedure in Melbourne,[2] we obtained data from 138 patients. Of these, only five possessed a ventricular mass made up of a solitary chamber. In the others, 84 had two ventricular chambers, but with the atrioventricular junctions arranged so that one of the chambers was obviously rudimentary and incomplete. In the remaining 49 patients, however, each atrial chamber was connected to its own ventricle, and some other morphologic anomaly had prevented the surgeon attempting a biventricular repair. Findings such as this are by no means unusual, and are virtually replicated in the experience from New Delhi. In their series of 240 patients undergoing a Fontan-type repair, only 3 possessed a truly solitary ventricle, considered by them to be of indeterminate morphology. Of the remaining 237, 101 had the atrial chambers connected predominantly to a big ventricle in the presence of a small and incomplete ventricular chamber, while the other 136 patients all had the arrangement in which each atrium was connected to its own ventricle.

It is gratifying for me as a morphologist to see that the team from New Delhi have found it possible to use the system of description which we developed to account for these complex malformations.[3,4] It has long been recognized that adequate description of hearts possessing one big and one small ventricle is difficult and contentious. The right ventricle in pulmonary atresia with intact ventricular septum, for example, is just as small, or even smaller, than the so-called "outlet chamber" in hearts with double inlet left ventricle, but possesses all its anatomic components whilst often being incapable of driving the pulmonary circulation. The outlet chamber in double inlet left ventricle, of course, is never capable of supporting the pulmonary circulation in its own right, but it used to be incorporated within the circulation feeding the lungs in some of the original modifications of the Fontan procedure. The anatomic solution to the problem in description is straightforward and simple - describe what is present. It is then justifiable to describe a functionally single ventricle in the situation where, anatomically, there is one big and one small ventricle. It no longer helps description, however, to attempt to deny ventricular status to the smaller chamber.

As the team from New Delhi explain so succinctly "The term single ventricle is anatomically incorrect, as two chambers can nearly always be seen in the ventricular mass, even though one ventricle is rudimentary and incomplete." The question now to be asked is why practitioners should still wish to call hearts "single ventricle" when they can be shown so clearly by modern-day imaging techniques to have

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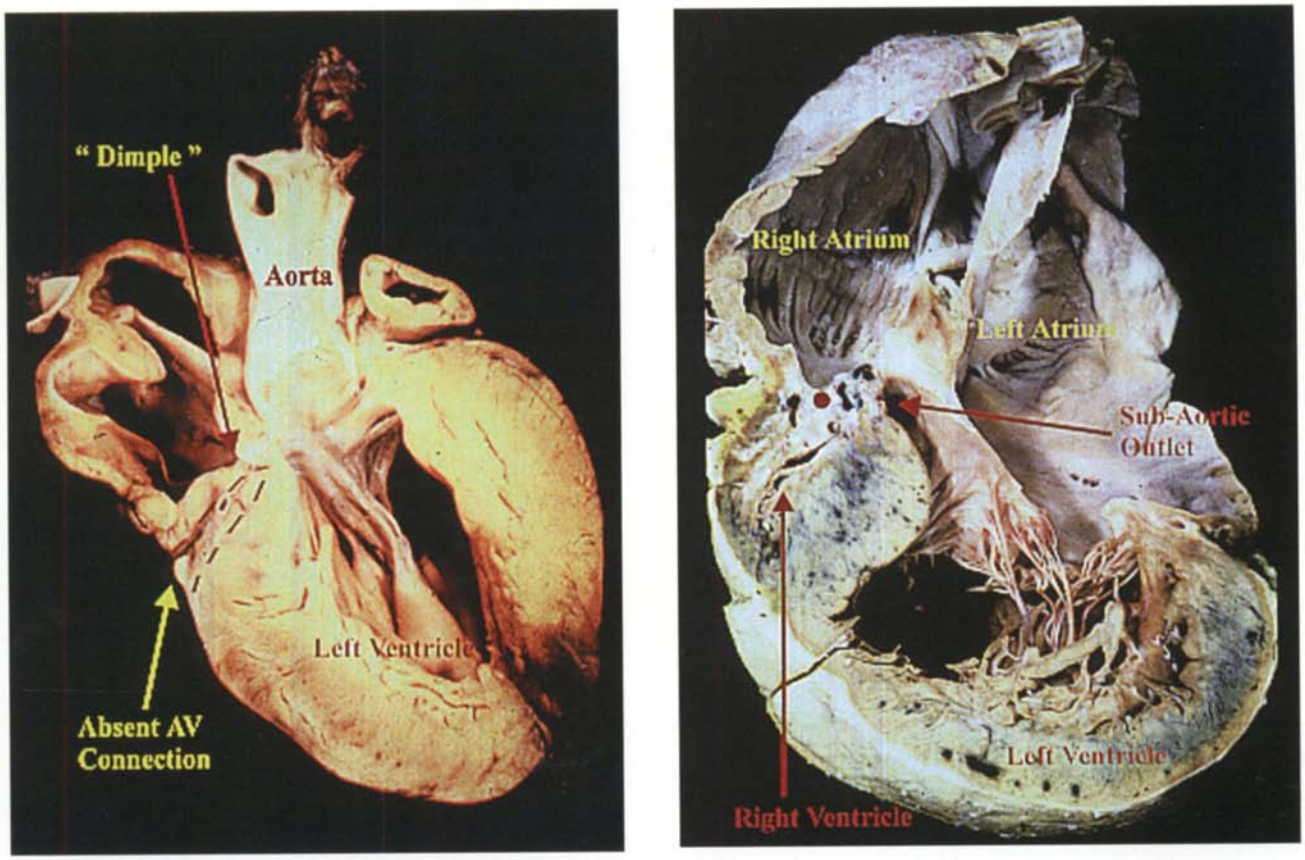


**Figure 1.**
*These cross-sections simulating the four-chamber echocardiographic images show the fundamental difference between absence of the right-sided atrioventricular (AV) connection (left hand panel) and an imperforate tricuspid valve (red dot) blocking the right atrioventricular connection in the setting of concordant atrioventricular connections (Right hand panel). Note that, in the heart with absent connection, by far the commonest substrate of "tricuspid atresia", the dimple points to the subaortic outflow tract. It represents the atrioventricular component of the membranous septum.*

one big and one small ventricle? It used to be conventional to use the term "single ventricle", or its synonym "univentricular heart", to distinguish hearts with double inlet left ventricle from those with the classical variant of tricuspid atresia. "Classical" tricuspid atresia is produced by absence of the right atrioventricular connection when the left atrium is connected to a dominant left ventricle. This distinction can now be made more readily and efficiently by describing the so-called "single ventricle" as double inlet left ventricle. There are still some who remain to be convinced that the typical variant of tricuspid atresia has absence of the right atrioventricular connection.[5] Should any still put currency in the concept that the echogenic mass seen echocardiographically represents an imperforate tricuspid valve, then they should compare anatomic examples with an imperforate membrane with those in which the atrioventricular groove interposes between the muscular floor of the right atrium and the base of the ventricular mass (Figure 1). Once the absent atrioventricular connection is recognized as a true anatomic entity, then it becomes crystal clear that the anatomic feature in common between tricuspid atresia and double inlet left ventricle is the fact that, in both lesions, only the dominant left ventricle is connected directly to the atrial chambers. This is because, in double inlet, both segments of atrial myocardium (right and left) are inserted into the dominant left ventricle whereas, in tricuspid atresia, as discussed, the right atrioventricular connection is completely absent (Figure 2).

Much of the trouble with use of "single ventricle", and "univentricular heart", stemmed from the comparison of hearts with double inlet left ventricle and tricuspid atresia. Controversies

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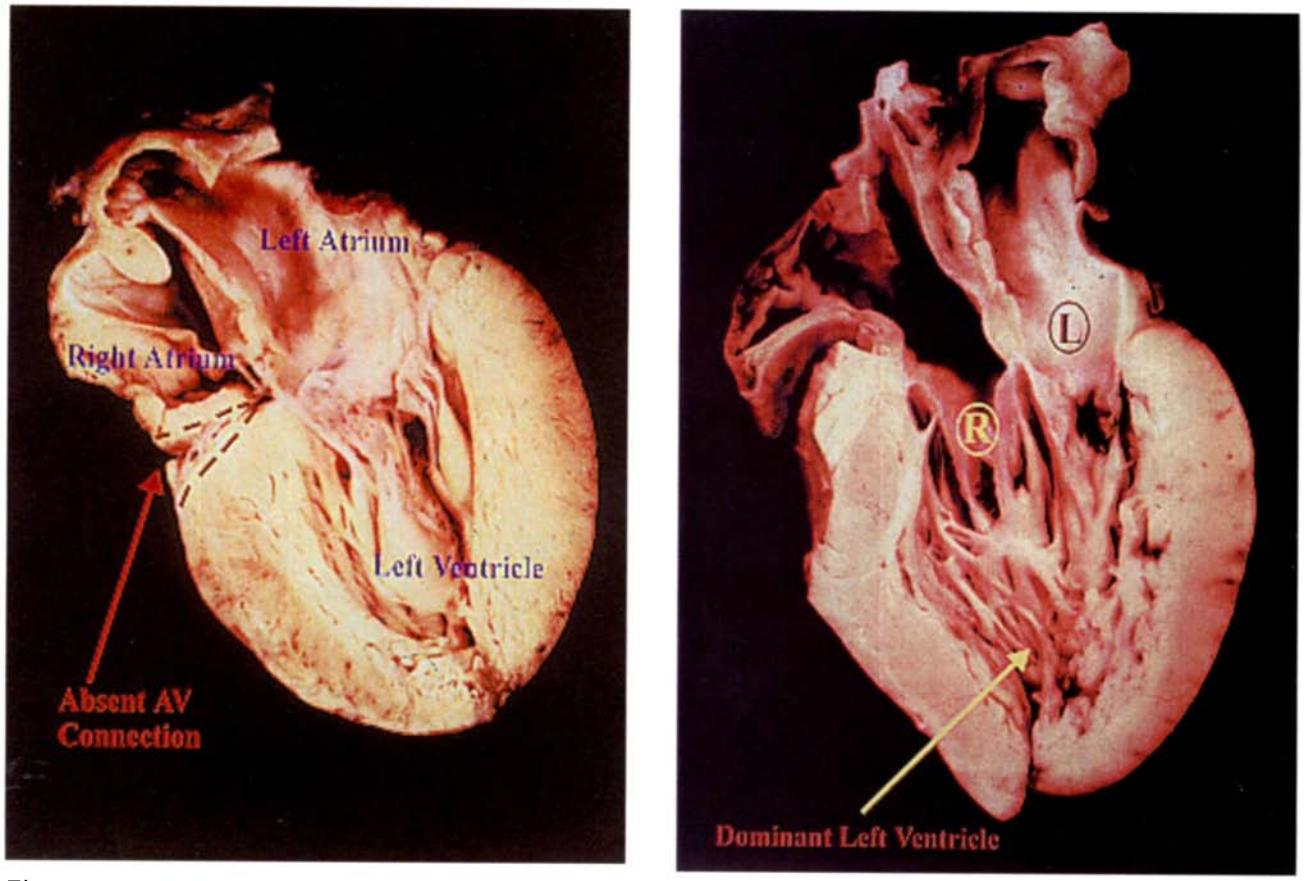


**Figure 2.**
*Further cross-sections simulating four-chamber echocardiographic images and demonstrating the similarities and differences between classical tricuspid atresia (left hand panel) and double inlet left ventricle (right hand panel). Both have a univentricular connection to a dominant left ventricle, but via only the left-sided atrioventricular (AV) connection in tricuspid atresia as opposed to right (R) and left (L) junctions in the setting of double inlet.*

raged during the 1970's and 1980's as to whether both were, or were not, univentricular. We had initially constructed formidable definitions so as to deny ventricular status to the incomplete right ventricular chamber in these hearts.[6] In this way, we attempted to justify inclusion of both entities in the univentricular category. We now recognize that this approach was ill-advised and illogical. It is the commitment of the atrioventricular junction in these entities which produces a univentricular situation, not the arrangement of the ventricular mass.[3,4] Underscoring all of the problems, however, was the old convention of dividing ventricles into "sinus" and "conus" components. There are no obvious landmarks for permitting this distinction in the normal heart. It is an easy matter, in contrast, to recognize that all ventricles possess inlets, apical trabecular components, and outlets. Normal ventricles have one of each. Abnormal ventricles can have more than one inlet or outlet, or can lack one or more of these components. The morphology of the ventricles, nonetheless, be they normal or abnormal, is determined according to the nature of the apical trabeculations. These can be of right ventricular, left ventricular, or solitary and indeterminate pattern (Figure 3). When ventricular structure is approached in this fashion, then even the most complex anomalies become straightforward in terms of description and categorization. As the group from New Delhi show so elegantly, analysis in this fashion sets the scene for the determination of the most appropriate surgical therapy.

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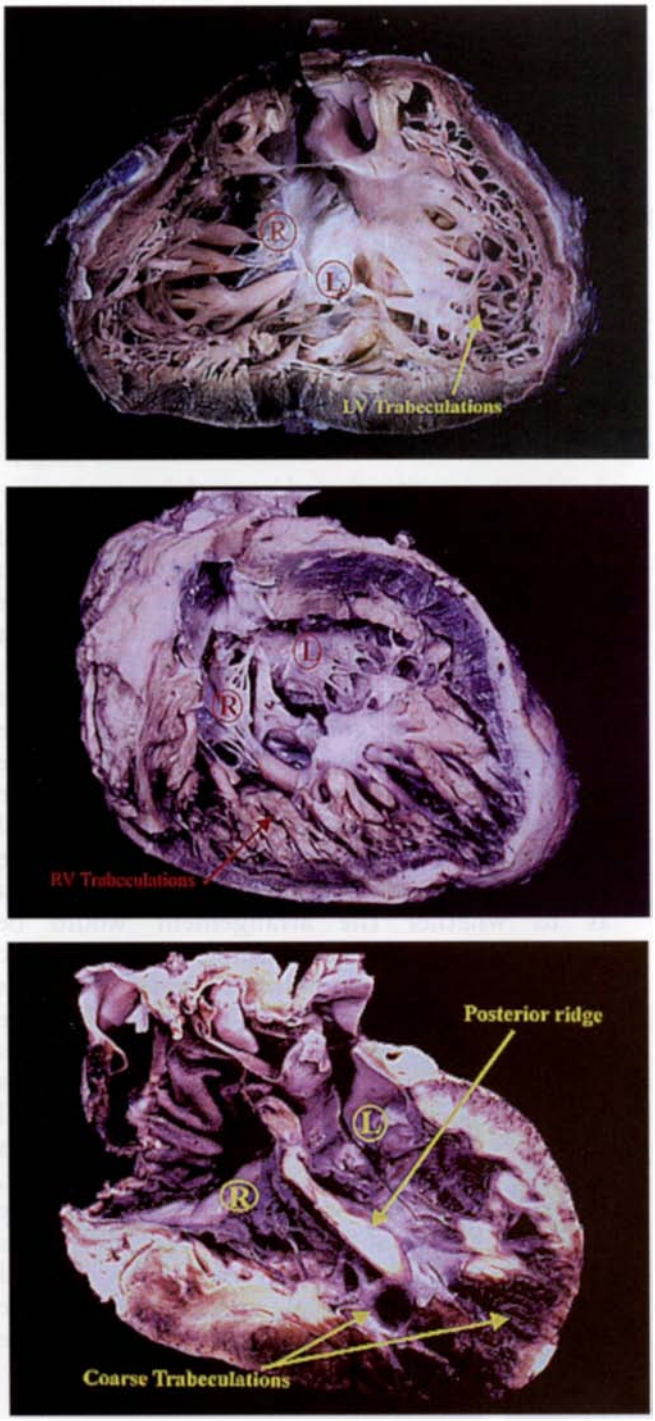


**Figure 3.**
*These dissections show how double inlet atrioventricular connection can be found with the atriums connected to a dominant left ventricle (LV - upper), a dominant right ventricle (RV - middle) or to a solitary and indeterminate ventricle (lower). The ventricles are distinguished morphologically according to the nature of their apical trabeculations, which show increasing coarseness from left, through right, to indeterminate ventricles. R - right atrioventricular junction; L - left atrioventricular junction.*

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